Cardiology in the Young

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Off-pump direct hepatic veins-to-hemiazygos vein anastomosis after primary Kawashima operation: long-term result

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Brief Report

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Abstract

Direct hepatic veins-to-hemiazygos connection offers the balanced distribution of hepatic venous blood to both lungs, not requiring anticoagulation. We report a 13-year follow-up after this type of off-pump Fontan completion. Patient's hepatic veins-to-hemiazygos confluence increased with growth to allow for unobstructed flow. This unique technique can be recommended in heterotaxy patients, if atrial hepatic venous drainage and hemiazygos vein are in close proximity.

Single-ventricle patients with interrupted inferior vena cava usually undergo staged Fontan surgery, starting with total cavopulmonary shunt (Kawashima operation). Months or years later, the hepatic veins get connected into the pulmonary circulation preventing the development or allowing for resolution of pulmonary arteriovenous malformations,[1] also eliminating desaturation originating from veno-venous collaterals. In Kawashima patients, almost always completion Fontan is established by the standard lateral or extracardiac tunnel.[2]

The direct hepatic veins-to-azygos/hemiazygos vein connection offers optimised streaming for hepatic venous blood completely mixing with systemic venous return. This approach had only been described in a few cases focusing on feasibility.[3–5] We presented long-term functional and anatomic results of direct hepatic veins-to-hemiazygos vein connection after Kawashima operation.

A 16-year-old girl with left atrial isomerism, single ventricle, d-malposition of the great arteries with pulmonary atresia and interrupted inferior vena cava with hemiazygos continuation to the left superior vena cava is now 13 years after completion Fontan surgery. There was no right-sided superior vena cava. After neonatal palliation with central aortopulmonary shunt, left-sided Kawashima operation was followed at 14 months leaving the hepatic venous return draining via intrapericardial inferior vena cava to right atrium. Almost 2 years later, she exhibited deteriorating physical activity and cyanosis; systemic arterial oxygen saturation decreased to 80%. Cardiac catheterisation/angiography revealed predominantly left-sided pulmonary arteriovenous malformations with desaturated left pulmonary venous blood (84%). It also revealed a close spatial relationship between the intracardiac inferior vena cava and the vena hemiazygos. This was verified by MRI (Fig 1).

The operation was performed at the age of 2 years and 11 months through a left posterolateral thoracotomy without cardiopulmonary bypass; a temporary conduit was interposed from the left-sided atrial appendage to the inferior vena cava before separating it from the right atrium. The intrapericardial inferior vena cava containing the hepatic venous drainage was directly anastomosed with the vena hemiazygos (enlarged with a small Gore-Tex patch). The post-operative course revealed transient intraoperative hepatic congestion, as seen by significant transaminase elevation and ascites, both peaking at 48 hours; at discharge on 15th post-operative day, percutaneous oxygen saturation was 90–95%. Low-dose oral acetylsalicylic acid was prescribed.

Our yearly follow-up protocol as in any Fontan surgery patient included abdominal sonography and liver function parameters, in addition, MRI and cardiac catheterisation early post-operatively and during long-term follow-up at 8 and 12 years.

Three months post-operatively (patient weight 12 kg, height 88 cm), cardiac catheterisation demonstrated a 4.5 mm narrowing at the hepatic vein-to-hemiazygos connection with a mean gradient of 2 mmHg (Fig 2a); Doppler sonography at the anastomosis revealed respiratory flow modulation with a maximum velocity of 0.7 m/s, but there was no signs of hepatic venous congestion and normal liver function tests.

Thirteen years later (patient weight 33 kg, height 147 cm), the diameter at the hepatic vein-to-hemiazygos connection had increased to 16 mm (Fig 2b) providing unobstructed hepatic venous drainage. Abdominal sonography showed mild heterogeneous echogenicity of the liver,

Figure 1. Preoperative MRI demonstrating the close proximity of the intrapericardial IVC (hepatic vein drainage black arrow) and hemiazygos vein (white arrowhead).

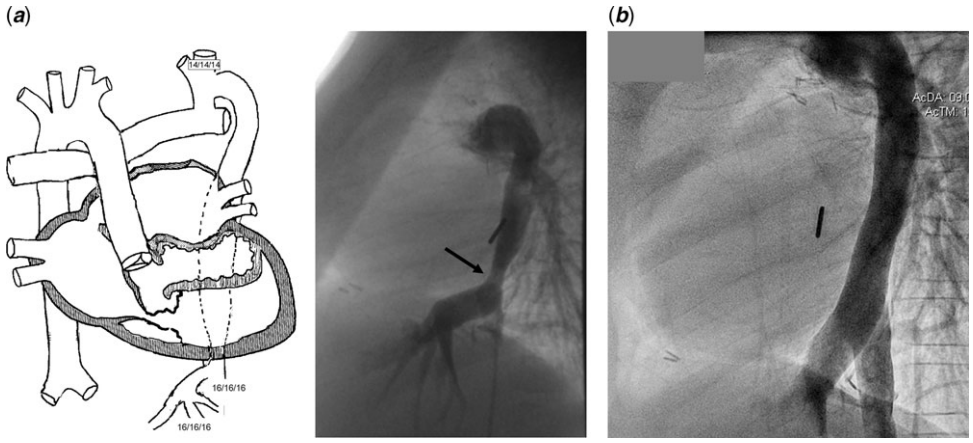

Figure 2. (***a***) Left: Post-operative schematic diagram showing the hepatic veins-to-hemiazygos connection. Right: Corresponding early post-operative angiography at 3 months revealed a mild narrowing immediately cranial to the hepatic veins-to-hemiazygos anastomosis within the hemiazygos (nearly 4–5mm) (black arrow). (***b***) Angiography after 12 years shows harmonious growth and unobstructed hepatic venous streaming/blood flow into the hemiazygos vein and farther up.

no surface abnormality/nodularity, or splenomegaly. Liver function tests revealed normal synthetic function and only slight elevation of the gamma-glutamyl transpeptidase.

Comment

Completion of the Fontan circulation in any stage of single-ventricle palliation, bringing hepatic venous blood directly into the pulmonary arteries, has shown to avoid and diminish existing pulmonary arteriovenous malformations, supporting the hypothesis of a hepatic factor in the pathogenesis of pulmonary arteriovenous malformations.[6] After total cavopulmonary shunt surgery (Kawashima operation) for univentricular heart with heterotaxy and interrupted inferior vena cava, completion Fontan employing the standard lateral tunnel or extracardiac conduit procedures creates risk for unilateral persistence of pulmonary arteriovenous malformations, due to imbalanced pulmonary flow distribution.[6,7]

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When carrying only hepatic venous flow instead of whole lower body return, standard lateral tunnel or extracardiac conduit procedure are at higher risk for thromboembolic complications. In addition, after a lateral tunnel procedure, atrial incisions and suture lines may predispose to atrial arrhythmia.[1,7] Lopez et al described the off-pump connection of the hepatic to azygos vein using a vascular prosthesis leaving the heart untouched.[5] As the risk for developing pulmonary arteriovenous malformations increases with follow-up, redirection of the hepatic venous blood is now recommended 6 months, at latest 2 years after Kawashima operation.[2]

In order to avoid all shortcomings of the standard lateral tunnel or extracardiac conduit procedures for Fontan completion in heterotaxy/interrupted inferior vena cava single-ventricle patients, especially assuring symmetric hepatic factor distribution, feasibility for direct intrapericardial inferior vena cava-azygos/hemiazygos vein anastomosis has to be evaluated. For this, MRI and cardiac catheterisation are essential to define the topography and the pattern of hepatic venous drainage.

In our patient, close proximity of the intrapericardial inferior vena cava (atrial hepatic venous drainage) to the hemiazygos vein was allowed for direct hepatic vein-to-hemiazygos vein anastomosis; extracorporeal circulation could be avoided using temporary tube drainage of hepatic venous flow to the left-sided atrial appendage. This still bears the risk of temporary hepatic venous congestion, as manipulations may distort the course of the tubes selected. Not only the diameter of the anastomosis itself, primarily enlarged with a small prosthetic patch, but also the narrowed hemiazygos vein at the anastomosis increased its diameter harmoniously with growth, and the angiography at follow-up examinations showed a "remodeling" of the anastomosis with laminar streaming. At the age of 16 years, unobstructed flow without a gradient from the hepatic veins-to-hemiazygos vein could be documented. The hemodynamic evaluation revealed mean pulmonary artery and hepatic vein pressure of 14 mmHg predisposing to changes in the liver structure – described as Fontan-associated liver disease in all patients with univentricular Fontan palliation.[8] Our patient, however, did not show any changes in liver function after 13 years.

While the feasibility of direct hepatic veins-to-hemiazygos vein connection to be performed as off-pump procedure has been shown before, we present the uncomplicated long-term follow-up for over 13 years. Besides the advantage of a balanced distribution of hepatic venous flow into the pulmonary arteries, avoidance of prosthetic interposition tube is less thrombogenic and also provides a potential for harmonious growth into adulthood without any obstruction.

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Conflicts of interest. None.

Ethical standards. This case report was performed according to the local code of research conduct and data protection rules and was according to the ethical guidelines and principles of the International Declaration of Helsinki. Approval by the local ethics committee was not required.

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